CASE REPORT

Transcatheter occlusion of a post-Fontan residual hepatic vein to pulmonary venous atrium communication using the Amplatzer septal occluder

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Abstract

A residual hepatic vein to left atrial communication may result in progressive cyanosis after the Fontan procedure. This problem has usually been treated surgically by ligation or re-inclusion of the residual hepatic vein in the Fontan circulation. Previous attempts at transcatheter closure of such veins have been unsuccessful. An Amplatzer septal occluder was successfully used for transcatheter closure of a post-Fontan hepatic vein to pulmonary venous atrium fistula in an 8 year old boy.

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Deliberate partial exclusion of a hepatic vein at the time of a modified Fontan procedure has been used as a form of fenestration.[1] Some patients may have previously unrecognised accessory hepatic veins draining to the left atrium.[2] Either arrangement may result in progressive cyanosis, as venous return is diverted through enlarging fistulae in the liver to the residual hepatic vein to pulmonary venous atrium connection because of the pressure gradient. The hepatic vein to atrial connection can be quite large and relatively short, making transcatheter occlusion difficult. This problem has usually been dealt with surgically by ligation or inclusion of the residual hepatic vein in the Fontan circuit. We describe a patient in whom a residual hepatic vein to atrial communication resulted in progressive cyanosis that was successfully treated with an Amplatzer septal occluder.

Case report

Our patient was born with right atrial isomerism, a common atrium, an atrioventricular septal defect with a small left ventricle, a double outlet right ventricle, and pulmonary stenosis. The right pulmonary veins drained into the common atrium and the left pulmonary veins drained into the innominate vein. The hepatic veins drained directly into the right atrium. He had a right sided modified Blalock-Taussig shunt at 3 months old. This was followed by a bidirectional superior cavopulmonary anastomosis and repair of partial anomalous pulmonary venous drainage at 27 months old. During this operation it was noted that one of the hepatic veins drained into the common atrium more to the left than usual. At age 67 months his Fontan circulation was completed by forming a lateral tunnel in the common atrium to baffle the inferior vena caval drainage and hepatic venous drainage to the pulmonary arteries. A 3 mm fenestration was created in the baffle. The previously described left sided hepatic vein could not be found. His postoperative recovery was uncomplicated and he was discharged home on the ninth postoperative day.

During outpatient follow up he became increasingly cyanosed with transcutaneous oxygen saturations falling from 87% at one year to 72% two years after the operation. Echocardiography showed that the fenestration was not particularly large and cardiac catheterisation was done when he was 8 years old.

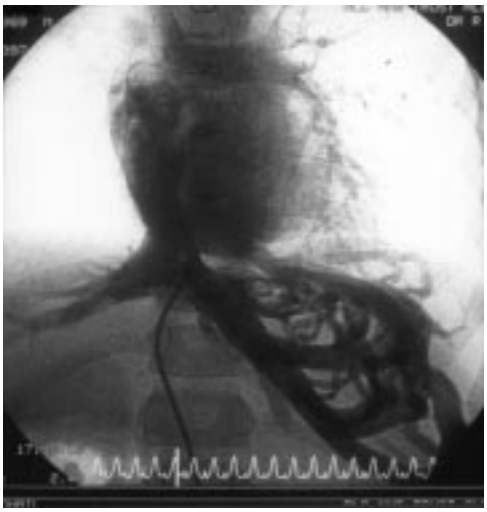

Figure 1 Angiogram of the inferior vena cava showing multiple venous collaterals in the liver draining into the residual left sided hepatic vein to pulmonary venous atrium fistula. The exit into the pulmonary venous atrium was 10 mm wide.

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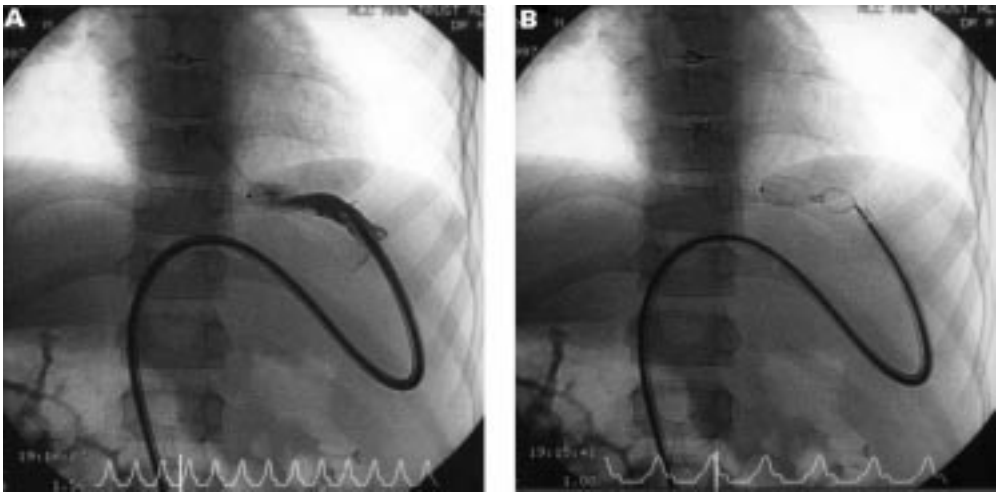

Figure 2 (A) A front loaded 10 mm Amplatzer septal occluder deployed at the exit of the fistula via a 7 F long sheath passed through a 10 F armoured long sheath. (B) The Amplatzer septal occluder has been released. An angiogram via the long sheath shows complete occlusion of the exit of the residual hepatic vein to pulmonary venous atrium fistula.

Under general anaesthesia arterial saturation was 86% and mean venous pressure 14 mm Hg. Angiography showed a small fenestration and multiple venous collaterals in the liver draining into the residual left sided hepatic vein (fig 1). The entry of this left sided hepatic vein into the pulmonary venous atrium was short and wide and appeared unsuitable for standard Gianturco coils or detachable balloons. We decided to use an Amplatzer septal occluder. This device was designed for occlusion of septal defects but can be compressed to plug tubular defects. It also has a very secure detachment mechanism that allows for easy retrieval or repositioning.

The residual left sided hepatic vein to atrial connection was entered initially via the fenestration and a stiff exchange wire was positioned in the hepatic vein. A 7 F long sheath was placed over the wire. However, the long sheath kinked at the fenestration once the dilator and wire had been withdrawn. The exit of the residual hepatic vein into the pulmonary venous atrium was then approached by passing a catheter from the hepatic veins and through the collaterals in the liver. A stiff exchange wire was then used to pass the 7 F long sheath. A 10 mm Amplatzer septal occluder was chosen, attached to the delivery wire, and compressed into the loader/introducer. The device was introduced into the long sheath and was easily advanced through the straight section of the sheath; however, considerable resistance was encountered when trying to advance the device through the curved part of the sheath's course. The device was withdrawn and a 10 F Arrow-flex armoured sheath substituted. Again the device could not be negotiated around the bend in the sheath. It was therefore loaded into the front end of a 7 F long sheath and the device and sheath assembly was easily advanced through the Arrow-flex sheath to the exit of the residual hepatic vein into the left atrium (fig 2A). The device was deployed by withdrawing the sheath(s). Angiography through the sidearm of the long sheath confirmed a satisfactory position with near complete occlusion. The device was then released by rotating the delivery cable anticlockwise. Angiography was performed again, which showed near complete occlusion (fig 2B). Arterial saturation rose to 98% and the central venous pressure and saturation remained unchanged.

Recovery was uncomplicated. The patient was given intravenous heparin by infusion until his INR rose above 1.8. Follow up echocardiography showed complete occlusion, and transcutaneous saturation measured at three months' follow up was 89%.

Discussion

The Amplatzer septal occluder was designed to occlude septal defects although its design origins relate to a triple disc vessel occluder made from a self expanding nitinol mesh.[3 4] This case demonstrates that the septal occluder design can be used to occlude large vessels. The device, when deployed, assumed an egg timer configuration because the discs of the device were compressed by the vessel walls; the device chosen was such that its central conjoined section was equal in diameter to the size of the vessel. A smaller diameter device could have been used provided there was sufficient outward pressure on the vessel to ensure device stability.

The size and lack of intrinsic stenoses in the exit of the fistula into the left atrium rendered it unsuitable for coil occlusion. Detachable balloons or coil sacs were another possibility but there are none large enough to block the fistula securely. A large, 17 mm diameter double umbrella or Cardioseal occluder could have been used to occlude this low pressure fistula. However, this device cannot be as easily withdrawn and repositioned before release.

Partial exclusion of a hepatic vein at the time of Fontan completion has been advocated as a method of fenestration to smooth postoperative recovery. The later occurrence of major intrahepatic venovenous fistulas resulting in severe progressive cyanosis has been well documented.[5 6] These have usually been treated surgically, either by ligating the fistula or by incorporation of the previously excluded

hepatic venous orifices by revising the previously placed baffle under hypothermic circulatory arrest. Transcatheter occlusion, to our knowledge, has not been successfully attempted before. Indeed, in one of the patients with accessory hepatic veins previously reported, a catheter passed through the fenestration was too unstable to permit coil occlusion and the patient died awaiting surgery. Our approach through the liver with a large reinforced flexible long sheath allowed a relatively large occlusion device to be delivered and its efficacy assured before detachment.

In conclusion, residual hepatic vein to left atria communications may result in progressive cyanosis after a Fontan procedure and can be successfully occluded by the Amplatzer septal occluder. However the device may need to be front loaded if the long sheath course is tortuous.

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